



Case review

Lethal aorto-oesophageal fistula – Characteristic features and aetiology

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ABSTRACT

Aorto-oesophageal fistula refers to a rarely encountered communication between the aorta and the oesophagus that results in massive and lethal haemorrhage into the upper gastrointestinal tract. Although the time between the initial haemorrhage and the terminal event may be days, a clinical diagnosis may not be established by the time of autopsy. A fistulous tract develops between the aorta and the oesophagus most commonly due to expansion of a thoracic aortic aneurysm or from an ingested foreign body. Less common causes include infiltrating neoplasms, oesophageal ulceration, vascular rings and iatrogenic lesions. Three cases are presented to illustrate the features of such cases due to aortic dissection, bronchial carcinoma and tuberculosis. Cases should be suspected if there has been a history of midthoracic pain or dysphagia, a 'herald' haemorrhage and then massive fatal haematemesis with bright red blood. Careful dissection at autopsy is required to demonstrate the site of the fistulous tract.

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1. Introduction

Aorto-oesophageal fistula refers to a communication between the aorta and the oesophagus that usually results in massive haemorrhage into the upper gastrointestinal tract with haematemesis, although rarely oesophageal contents may find their way into the circulation.¹ It is an uncommon event but one which often has a rapidly lethal outcome. As this may result in the need for a forensic autopsy, the characteristic features of this condition and the possible causes have been reviewed to provide an overview of the clinical presentation and an analysis of the range of possible initiating events and conditions.

2. Materials and methods

Case files at The University of Adelaide Discipline of Anatomy and Pathology were reviewed for illustrative examples of aorto-oesophageal fistula. Case details were summarised and images obtained. A literature search was also performed of the United States National Library of Medicine 'Entrez PubMed' database (<http://www.ncbi.nlm.nih.gov/pubmed>) for all entries listed under 'aorto-oesophageal fistula'.

3. Results

Three cases were selected to demonstrate different underlying aetiological processes.

Case 1: A 77-year-old man with a history of previous aortic dissection presented to hospital with haematemesis and melaena. An upper gastrointestinal endoscopy revealed blood clot within the oesophagus and an aorto-oesophageal fistula was diagnosed. This was followed soon after by further haemorrhage and death. At autopsy the major findings were cardiovascular with an oval defect in the descending thoracic aorta which communicated with an old dissection that extended to the left common iliac artery. The defect also communicated with an 80-mm-diameter saccular pseudoaneurysm that had eroded into the adjacent oesophagus. The defect in the oesophagus measured 45 mm in maximum dimensions and was filled with a 72-g blood clot (Fig. 1). The stomach and proximal small intestine contained 1.8 l of fresh blood. Death was due to haemorrhage from an aorto-oesophageal fistula arising from a pseudoaneurysm in a previous aortic dissection.

Case 2: A 59-year-old woman with a poorly differentiated carcinoma of the upper lobe of the left lung treated with radiotherapy died soon after presentation to hospital with haemoptysis, haematemesis and melaena. At autopsy, a fistula tract was demonstrated extending from an ulcerated defect in the descending portion of the thoracic aorta (Fig. 2), through a mass of necrotic

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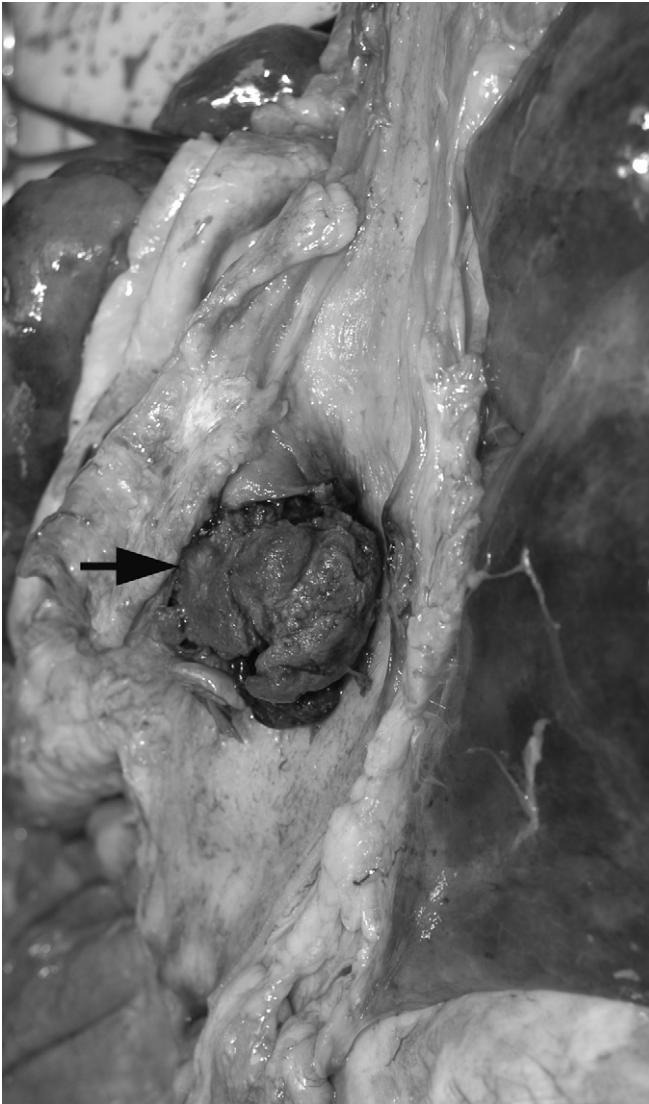


Fig. 1. Ulceration of the oesophagus with the defect plugged by a blood clot (arrow) that extends into an underlying pseudoaneurysm of the aorta in case 1.

metastatic tumour within mediastinal lymph nodes to an ulcerated defect in the oesophagus. The stomach was distended and full of blood clot, with fresh blood within both the small and large intestines. A round tumour mass was present in the upper lobe of the left lung and the trachea was ulcerated by infiltrating tumour within paratracheal lymph nodes. Death was due to haemorrhage from an aorto-oesophageal fistula arising within an infiltrating metastatic lung tumour.

Case 3: A 42-year-old man with a history of active tuberculosis presented to hospital with bright red haematemesis. There was no history of peptic ulcer or liver disease. Further massive bleeding occurred which failed to respond to treatment and death occurred shortly thereafter. At autopsy, the oesophagus was adherent to a large mass of necrotic tuberculous material. A 2-cm ulcer led into a fistula tract that extended through the necrotic material to the aorta which was also adherent to the mass. A 2-cm full-thickness defect in the aortic wall communicated with the tract (Fig. 3A and B). Further tuberculous lesions were identified in the lungs and seminal vesicles. Death was due to haemorrhage from an aorto-oesophageal fistula arising within a mass of necrotic tuberculous mediastinal lymph nodes.



Fig. 2. Ulceration of the aorta (arrow) due to infiltration by a metastatic tumour deposit in mediastinal lymph nodes from a primary poorly-differentiated carcinoma of the lung in case 2.

4. Discussion

Aorto-oesophageal fistula, which is characterised by Chiari's triad of midthoracic pain or dysphagia, followed by a 'herald' haemorrhage in 80% of cases, and then massive fatal haematemesis, was first described in the early nineteenth century.¹ Although the time between the initial haemorrhage and the terminal event may be days, a clinical diagnosis may not have been established,² placing the responsibility for the diagnosis firmly with the forensic pathologist. Pain is caused by distension or dissection of the aortic wall, or by oesophageal leakage with mediastinitis.² The bleeding is often bright red, in contrast to venous bleeding from oesophageal varices. Aortoenteric fistulae are responsible for only 3.5% of cases of upper gastrointestinal haemorrhage, with the majority located between the aorta and the duodenum.¹ The types of lesions and conditions that may cause a communication to develop specifically between the aorta and the oesophagus are quite varied ranging from trauma and ulceration, to infection and neoplasia, and are summarised in Table 1.

The most common cause of a primary aorto-oesophageal fistula is an atherosclerotic aneurysm of the thoracic aorta (in 51–75% of cases).^{3,4} Gradual enlargement of a thoracic aneurysm results in pressure necrosis of the adjacent oesophagus with inflammation and adhesion formation, and subsequent thinning of the aneurysm

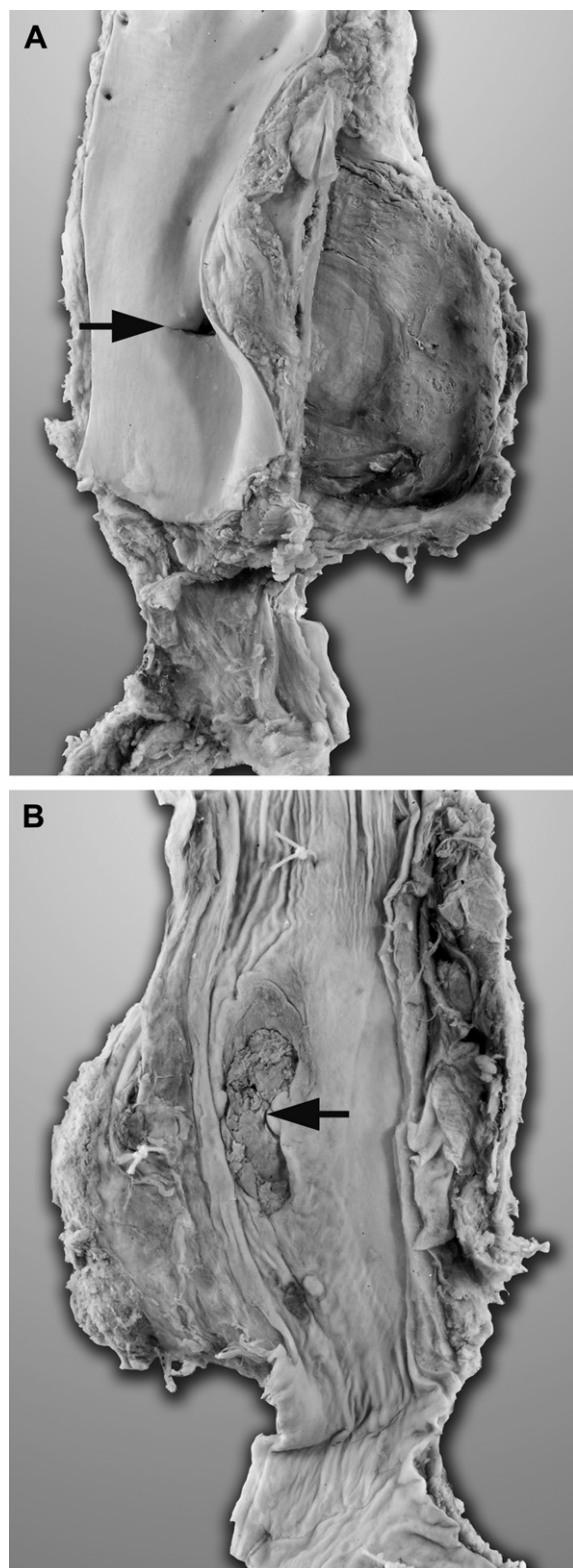


Fig. 3. Opened aorta showing an ulcerated defect due to erosion by a large mass of necrotic tuberculous tissue in case 3 (arrow) (A). The same specimen viewed from the oesophageal side showing the full thickness defect in the oesophageal wall communicating with the fistula tract (arrow) (B).

wall with eventual rupture.^{2,5} This usually occurs in the descending aorta where the aorta and oesophagus are in close proximity; however, cases have occurred at all parts of the thoracic aorta.¹ Following surgical resection of an aneurysm, anastomotic

Table 1

Causes of lethal aorto-oesophageal fistula.

Aortic aneurysm
i) Atherosclerotic
ii) Dissecting
iii) Syphilitic
Infiltrating neoplasm
i) Oesophagus
ii) Lung
iii) Mediastinal
Infection
i) Tuberculosis
ii) Mycotic aneurysm
iii) Mediastinal abscess
Penetrating oesophageal ulcer
Aortitis
Congenital vascular anomaly
Swallowed foreign body
Ingested corrosive material
Chest trauma
i) Blunt
ii) Sharp/penetrating
Iatrogenic
i) Post surgical repair
a) Aortic
b) Oesophageal
ii) Oesophageal instrumentation
iii) Aortic cannulation
iv) Radiotherapy

disruption may also lead to the development of a fistula.⁶ Less common types of fistulae arise from dissecting aortic aneurysms where blood has tracked down within the layers of the aortic wall. Case 1 illustrates this situation, with a delayed rupture of a pseudo-aneurysm formed by expansion of the outer layers of a dissected thoracic aorta, with eventual rupture into the oesophagus. A 'herald' haemorrhage had occurred in this case resulting in both haematemesis and melaena. Bleeding had most likely been stopped by plugging of the defect with blood clot, as could be seen at autopsy (Fig. 1). Bacterial infection, or the corrosive action of gastrointestinal contents, may weaken the clot resulting in further haemorrhage.² Other causes of cessation of the initial haemorrhage include arterial spasm and hypotension due to blood loss.² Syphilitic aneurysms are less common nowadays and rarely aortitis may initiate the formation of a fistula.^{7,8}

Malignant neoplasms may compromise aortic and oesophageal integrity. The most common tumours that result in aorto-oesophageal fistula are carcinomas of the oesophagus and bronchi, although any infiltrating mediastinal mass has the potential to cause a fistula. Oesophageal carcinomas are a particular risk, as 87% arise in the middle third of the oesophagus where it is closely apposed to the aorta. Carcinomatous ulcers have perforated the oesophagus in as many as 45% of cases. Other vessels that may be eroded by oesophageal tumours include the carotid, subclavian, oesophageal and intercostal arteries.¹ An example of a bronchogenic carcinoma spreading into the space between the oesophagus and aorta and eroding both is shown in case 2. Rarely metastatic tumours from sites as far removed as the cervix may lodge in the oesophagus and erode the adjacent aorta.¹ On occasion, tumour infiltration into the vasculature will not cause haemorrhage, but will allow passage of upper gastrointestinal bacteria into the circulation with recurrent septicaemias/bacteraemias that may be incorrectly attributed to bacterial endocarditis.⁹

The mechanism by which infiltrating carcinomas lead to aortic perforation has been debated, with evidence that it does not

necessarily rely on pressure necrosis or bacterial infection, as penetration of the aortic wall is often only to the level of the adventitia. Instead, it is proposed that necrosis of the aortic wall is induced by compromise of blood flow through the vasa vasorum because of thrombosis.¹

Infections within the mediastinum may have a similar effect on neoplasms with erosion of the aortic and oesophageal walls providing a conduit between the two. Tuberculosis may weaken the aortic wall either by primary tuberculous aortitis or by local tuberculous abscess formation.¹ In case 3 an active tuberculous infection can be seen adherent to both the aorta and oesophagus (Fig. 3). Any infective cause of mediastinal abscess or mycotic aneurysm could potentially have this effect.

Erosive oesophagitis with ulceration may result in perforation through the oesophageal wall into the aorta.⁴ On occasion, persistent reflux of acid gastric contents into the lower oesophagus may cause a Barrett's oesophagus where the distal oesophagus is lined by columnar epithelium in continuity with the gastric mucosa. Barrett's oesophagus is associated with ulceration and perforation and may result in lethal aorto-oesophageal or oesophagopleural fistulae.^{10–12}

Abnormally positioned vessels forming vascular rings may be associated with aorto-oesophageal fistula formation.¹³ An example of this is congenital double aortic arch, where the oesophagus, compressed by an abnormally placed aorta, may be eroded by prolonged nasogastric intubation.¹⁴ Erosion of the wall of the oesophagus by a suture from a previously ligated patent ductus arteriosus has been reported in a 4-year-old girl who died following a massive haematemesis.¹³

Foreign bodies are the second most common cause of aorto-oesophageal fistulae, being responsible for 19% of cases (although conversely less than 0.1% of impacted foreign bodies cause fistulae).³ Foreign-body perforation may result in local sepsis with abscess formation at any level of the gastrointestinal tract¹⁵ but in the oesophagus it most often occurs at the point where the oesophagus is narrowed by the aortic arch.² The nature of the foreign body varies with age, with adults most often having problems with ingested animal bones (e.g., fish and chicken) or dental prostheses, and children with coins, pins and needles.^{3,16,17} Eighty percent of foreign-body ingestion occurs in children, most under 3 years of age.¹⁸ A large number of ingested foreign bodies, with materials such as broken glass, suggest an underlying psychiatric illness. Sharp foreign bodies such as bone shards may perforate the wall directly, while others may impact and erode by pressure necrosis.¹⁹ This may be exacerbated by objects such as button batteries which contain corrosive material such as sodium or potassium hydroxide.^{13,18} Foreign bodies may initially cause mediastinitis or aortitis which then secondarily erodes vascular walls.² In addition to creating fistulae between the oesophagus and aorta, impacted coins may erode towards other major vessels, such as the common carotid artery.²⁰ Survival after foreign-body-induced aorto-oesophageal fistula is rare with only seven out of 100 cases in the literature being alive after 12 months.³ The ingestion of corrosive material such as lye may also cause significant erosive oesophagitis with perforation of the underlying oesophageal wall.¹

Chest trauma from either blunt impact or a penetrating injury may lead to the development of a false aneurysm of the thoracic aorta which may then progress to fistula formation as it enlarges.¹

Iatrogenic causes of aorto-oesophageal fistula include instances where there has been tissue breakdown after aortic or oesophageal surgery. Surgical treatment of thoracic aortic aneurysms may be by open repair or by endovascular stenting (thoracic endovascular aortic repair or TEVAR), both of which have been associated

with the development of secondary aorto-oesophageal fistulae. Fistulae are believed to develop from chronic pulsation of the prosthetic graft against the oesophagus, or from a low-grade infection of the graft with the development of a pseudo-aneurysm.^{4,21} TEVAR may also lead to aortobronchial fistulae with lethal haemoptysis.⁵

Oesophagectomy, usually for oesophageal carcinoma, has been associated with the formation of secondary aorto-oesophageal fistulae due mainly to a failure of the intrathoracic anastomosis. Leakage of oesophageal and gastric contents results in mediastinal abscess formation.²² In an attempt to avoid this complication, endoscopic stent repair was introduced; however, this may also result in aorto-oesophageal fistulae due to pressure on the adjacent aorta with necrosis of the vessel wall.^{22,23} Less commonly, erosions or sinuses from grafts may extend out from the oesophagus without communicating with the lumen of the aorta and so do not cause massive haemorrhage; the presentation instead involves mediastinitis and graft infection.⁶ Other iatrogenic causes of fistulae include oesophageal instrumentation and endovascular catheters in the aorta resulting in tears with pseudo-aneurysm formation, and radiotherapy for tumours that induces aortic necrosis.¹

Although most cases of lethal upper gastrointestinal haemorrhage involve peptic ulceration, oesophageal varices and Mallory–Weiss tears, occasional cases may arise from aorto-oesophageal fistulae. A characteristic history with a 'herald' bleed followed by bright red haematemesis may provide a hint as to the diagnosis prior to autopsy. Careful dissection of the mid-thoracic region will be required to demonstrate the exact location of the fistula and to elucidate the precise nature of the particular predisposing condition.

Conflict of interest

None.

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Ethical approval

N/A.

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